

SHOEMAKER (G.E.)

A CASE OF CONGENITAL DEFICIENCY OF THE LOWER EXTREMITIES.

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BY
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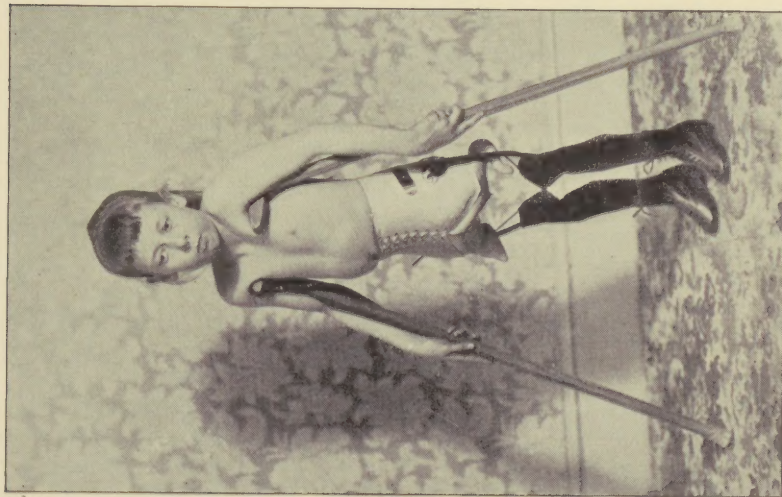


FIG. 1.

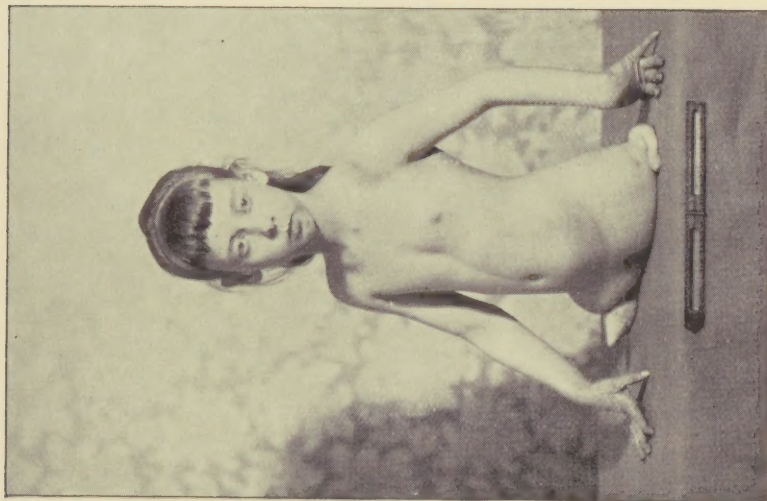


FIG. 2.—Sitting position.



FIG. 3.—Shows position assumed when walking on the hands.

A CASE OF CONGENITAL DEFICIENCY OF THE LOWER EXTREMITIES.

THE child before us was born and now lives in Maine. She was recently sent to this city and admitted to the writer's wards at the Methodist Hospital for the purpose of securing mechanical support. She is shown here simply as a congenital deformity, though it may be said that since coming here, a few weeks since, she has learned to walk readily by the aid of crutches,—first upon rigid rods, forked at the top and attached to the body by a plaster-of-Paris cup enveloping the pelvis. The weight of the body rested entirely upon the tuberosities of the ischia, as in the normal sitting posture. The apparatus shown in the cut is simply an elaboration of the same idea: a pelvic cup, with two rigid metallic limbs fastened together and jointed at the sides of the pelvis. The joint has a simple catch, or lock, which can be loosened by the hand when the girl wishes to sit down. The pelvic part is of rawhide, moulded over a cast of the patient and finished in the same manner as the spinal jackets made by Mr. Gemrig, of this city. The sole of the shoe is rigidly attached to a thin steel plate which answers for a foot. The stockings are lightly padded with wool. The child goes about with the apparatus with great facility and entire comfort. Owing to the activity and tender age of the child, it is not considered advisable to complicate the apparatus by any knee-joints, though were she an adult a fairly useful joint might be devised for that situation.

The child is nine years of age, and is one of seven living children, none of the others presenting deformities. The parents are living and normal.

From the stand-point of teratology the case is of much interest. The deformity of the lower limbs alone is said by Geoffroy-Saint-Hilaire to be more rare than that of the upper, and also more rare than a deficiency in all four. In Ahlfeld's work on deformities I was unable to find a case of precisely the same type. The recent work of Dr. Hirst and Dr. Piersol does not do more than refer to a classification in the folios so far issued. However, M. Breschet (*Bulletin de la Faculté de Médecine*, t. iv. p. 328) describes deformities in an adult male curiously similar to the present case. The man was at that time—early in the present century—a great travelling curiosity. In his case and in this the rudiments are very unlike on the two

sides, but, curiously enough, the conditions present are exactly reversed as far as the sides are concerned.

One may recall the general division of members which is observed in man and many higher vertebrates into four segments: first, the one nearest the body, for the attachment of the member to the trunk, illustrated in man by the shoulder-girdle or half the pelvis; second and third, the intermediate segments, as the arm and forearm, or the thigh and leg; finally, the end segment, represented by the hand or foot.

In the case before us the first or pelvic segment remains apparently normal in bony development, with no evidence of abnormal origin of muscles. For instance, the glutei are well developed, as are apparently the psoas and iliacus and the upper ends of muscles arising from the front of the ilium which would flex the thigh.

However, the insertions of these muscles into the rudiments of extremities are obscure, as the bony elements of the rudiments are so small and so abnormal in shape that no certainty attaches to any attempt at identification.

The difficulty is at once understood when it is stated that both rudiments are buried within the contour of the buttock to a considerable degree. No hip-joint is present on either side, and probably no femur, though the structure of the left rudiment is less clear than that of the right.

In the right rudiment the soft parts resemble to some extent a small leg and foot, but the resemblance is entirely superficial. Instead of the normal thirty bones, the limb below the pelvis appears to contain only two. There is no hip-joint and no femur. There is one long bone, probably the tibia, which has a musculo-ligamentary attachment to the pelvis in front of and below the anterior superior spinous process of the ilium where it is buried beneath the integument and tissues of the trunk. This bone is about five inches in length, and extends downward and forward to the very tip of the single toe-like extremity. There is no nail upon this tip, but the skin is hardened by use. The other bone is short and irregular, not over an inch in length, and is felt behind and a little above the lower end of the longer bone. It is freely movable, gives substance to the cushion used as a heel, and is probably a rudimentary fibula.

The tarsus, metatarsus, and phalanges are completely absent. In other words, the only portion of the limb present on this side is an imperfect lower intermediary segment. There is no true joint present, and the free, voluntary movements in all directions take place through the muscular attachment of the single long bone to the pelvis. The limb may be elongated about two inches by traction downward, the upper end of the long bone sliding along the pelvis. It still has a large degree of strength, and the girl is able to stand unsteadily upon the two rudiments, when the buttocks are raised so as to clear the floor about half an inch.

The left extremity is much weaker and more limited in motion than the right, though it contains more parts. The foot segment is represented by a single well-formed toe, with its metatarsal bone. A toe-nail of ordinary

appearance is observed. There is no tarsus. A rudimentary tibia and fibula, about four and three-quarters inches long and very thin and weak, may be traced partly within the buttock. From the upper end of the tibia a stronger mass extends backward towards the acetabulum, but its identity as a femur is uncertain because of its being buried in the buttock. The whole limb is flexed or folded upon itself till, in standing, the finger-like foot lies upon the floor and the fibula and tibia lie almost horizontally upon it, and are in contact with it for a considerable distance. This throws what should be the knee in front of and below the anterior superior process of the ilium, but it is completely covered by the tissues of the anterior part of the buttock or trunk.

The limb cannot be straightened to any considerable degree, and its chief motion is backward, though that is quite limited. There are possibly present some elements of every segment of a normal limb, though no positive evidence of a femur exists.

There are no other deformities in other parts of the body. The large development of the shoulders and upper arms is due to over-use. The elbow-joint extends over about an inch and a half beyond a straight line, as though from its unwonted strain. The triceps on each side is better developed than the biceps, because she uses her arms as legs in walking. The biceps is assisted in maintaining rigid extension by the chocking of the elbow-joint. The triceps is not so assisted and does more work. The genitalia and anus appear normal, though the labia majora are rather small. It is usual for deformed individuals of this kind to be sexually deficient, though the man to whom allusion was made as resembling this case was the father of several children.

This child is strong and active, and has always been well. The height when sitting is twenty-seven inches, which is increased to twenty-nine inches when she "stands" resting on the rudiments and balancing with the hands upon the floor. In this position the weight is borne on the last phalanges of the thumbs and on the metacarpo-phalangeal joints. The chest at the nipple-line measures twenty-three and a half inches after expiration, and twenty-five and a half inches after inspiration; waist at umbilicus, twenty and a half inches; circumference of hips, twenty-four and a half inches. The general development corresponds to that of the average girl at her age.

She walks at times by using her arms like crutches and swinging the body forward between them. She also walks on the stumps, or, more properly, on the buttocks, swinging first one and then the other forward. She walks readily upon her hands, with the pelvis high in the air, as shown in the photograph. Her intelligence is good, though she has been much repressed, her family being ignorant. She has no education, though that is not for want of capacity.

No perfectly satisfactory explanation can be given for the failure in development. It may simply be said that any theory of intra-uterine amputation or constriction is, to my mind, disproved, at least on the left side, by the persistence of the single well-formed toe and its metatarsal bone, with

the complete absence of traces of the other toes. Abnormal distribution of blood-supply would not account for this, and we are thrown back upon trophic influences, probably of central origin, whose nature in the last analysis is unknown.

It may be recalled that there is a classification of monstrosities which depends upon the development of the limbs. Thus, a monster without limbs belongs to the class *Amelus* (μέλος, *a limb*). One with two limbs fused together becomes *Symelus*. The *Phocemelus* has a terminal segment of a limb—for instance, the hand—attached directly to the shoulder-girdle without the intervention of the intermediary segments, as the arm and forearm. This arrangement is normal in some marine animals, hence the name *Phocemelus* (φώκη, *a seal*; μέλος, *limb*). In the *Hemimelus* two distal segments are wanting. It will readily be seen that the present case belongs to none of these groups, but must be classed as *Ectromelus* (ἐκτρομωσις, *abortion*), according to the classification of Geoffroy-Saint-Hilaire and others, there being simply a failure in development. The accompanying illustrations (opposite page 136) are from photographs.

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EDITED BY JUDSON DALAND, M.D.,

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